RAW SEQUENCE LISTING

The Biotechnology Systems Branch of the Scientific and Technical Information Center (STIC) no errors detected.

Application Serial Number: /0/53/,427
Source: /FWP
Date Processed by STIC: 8/4/06

ENTERED



IFWP

RAW SEQUENCE LISTING DATE: 08/04/2006 PATENT APPLICATION: US/10/531,427 TIME: 13:11:47

Input Set : A:\2005-11-10 0020-5363PUS1.txt
Output Set: N:\CRF4\08042006\J531427.raw

```
3 <110> APPLICANT: MIYATA, Toshiyuki
             KOKAME, Koichi
     6 <120> TITLE OF INVENTION: SUBSTRATES SPECIFIC TO VON WILLEBRAND FACTOR CLEAVING
PROTEASE AND METHOD
             OF ASSAYING THE ACTIVITY
     9 <130> FILE REFERENCE: 0020-5363PUS1
    11 <140> CURRENT APPLICATION NUMBER: US 10/531,427
    12 <141> CURRENT FILING DATE: 2005-04-15
    14 <150> PRIOR APPLICATION NUMBER: PCT/JP02/10816
    15 <151> PRIOR FILING DATE: 2002-10-18
    17 <160> NUMBER OF SEQ ID NOS: 12
    19 <210> SEQ ID NO: 1
    20 <211> LENGTH: 2813
    21 <212> TYPE: PRT
    22 <213> ORGANISM: Homo sapiens
    24 <400> SEQUENCE: 1
    25 Met Ile Pro Ala Arg Phe Ala Gly Val Leu Leu Ala Leu Ala Leu Ile
    27 Leu Pro Gly Thr Leu Cys Ala Glu Gly Thr Arg Gly Arg Ser Ser Thr
                    20
    29 Ala Arg Cys Ser Leu Phe Gly Ser Asp Phe Val Asn Thr Phe Asp Gly
    31 Ser Met Tyr Ser Phe Ala Gly Tyr Cys Ser Tyr Leu Leu Ala Gly Gly
    33 Cys Gln Lys Arg Ser Phe Ser Ile Ile Gly Asp Phe Gln Asn Gly Lys
    35 Arg Val Ser Leu Ser Val Tyr Leu Gly Glu Phe Phe Asp Ile His Leu
    37 Phe Val Asn Gly Thr Val Thr Gln Gly Asp Gln Arg Val Ser Met Pro
                   100
                                        105
                                                            110
    39 Tyr Ala Ser Lys Gly Leu Tyr Leu Glu Thr Glu Ala Gly Tyr Tyr Lys
                                    120
    41 Leu Ser Gly Glu Ala Tyr Gly Phe Val Ala Arg Ile Asp Gly Ser Gly
                                135
    43 Asn Phe Gln Val Leu Leu Ser Asp Arg Tyr Phe Asn Lys Thr Cys Gly
    45 Leu Cys Gly Asn Phe Asn Ile Phe Ala Glu Asp Asp Phe Met Thr Gln
    46
                       165
                                            170
    47 Glu Gly Thr Leu Thr Ser Asp Pro Tyr Asp Phe Ala Asn Ser Trp Ala
                                        185
    49 Leu Ser Ser Gly Glu Gln Trp Cys Glu Arg Ala Ser Pro Pro Ser Ser
               195
                                   200
    51 Ser Cys Asn Ile Ser Ser Gly Glu Met Gln Lys Gly Leu Trp Glu Gln
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220

215

210

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53 Cys Gln Leu Leu Lys Ser Thr Ser Val Phe Ala Arg Cys His Pro Leu 54 225 230 55 Val Asp Pro Glu Pro Phe Val Ala Leu Cys Glu Lys Thr Leu Cys Glu 250 57 Cys Ala Gly Gly Leu Glu Cys Ala Cys Pro Ala Leu Leu Glu Tyr Ala 260 265 59 Arg Thr Cys Ala Gln Glu Gly Met Val Leu Tyr Gly Trp Thr Asp His 275 280 61 Ser Ala Cys Ser Pro Val Cys Pro Ala Gly Met Glu Tyr Arg Gln Cys 295 63 Val Ser Pro Cys Ala Arg Thr Cys Gln Ser Leu His Ile Asn Glu Met 310 315 65 Cys Gln Glu Arg Cys Val Asp Gly Cys Ser Cys Pro Glu Gly Gln Leu 325 330 67 Leu Asp Glu Gly Leu Cys Val Glu Ser Thr Glu Cys Pro Cys Val His 340 345 69 Ser Gly Lys Arg Tyr Pro Pro Gly Thr Ser Leu Ser Arg Asp Cys Asn 360 71 Thr Cys Ile Cys Arg Asn Ser Gln Trp Ile Cys Ser Asn Glu Glu Cys 370 375 380 73 Pro Gly Glu Cys Leu Val Thr Gly Gln Ser His Phe Lys Ser Phe Asp 390 395 75 Asn Arg Tyr Phe Thr Phe Ser Gly Ile Cys Gln Tyr Leu Leu Ala Arg 405 410 77 Asp Cys Gln Asp His Ser Phe Ser Ile Val Ile Glu Thr Val Gln Cys 420 425 430 79 Ala Asp Asp Arg Asp Ala Val Cys Thr Arg Ser Val Thr Val Arg Leu 435 440 81 Pro Gly Leu His Asn Ser Leu Val Lys Leu Lys His Gly Ala Gly Val 455 83 Ala Met Asp Gly Gln Asp Val Gln Leu Pro Leu Leu Lys Gly Asp Leu 470 85 Arg Ile Gln His Thr Val Thr Ala Ser Val Arg Leu Ser Tyr Gly Glu 485 490 87 Asp Leu Gln Met Asp Trp Asp Gly Arg Gly Arg Leu Leu Val Lys Leu 500 505 89 Ser Pro Val Tyr Ala Gly Lys Thr Cys Gly Leu Cys Gly Asn Tyr Asn 515 520 91 Gly Asn Gln Gly Asp Asp Phe Leu Thr Pro Ser Gly Leu Ala Glu Pro 535 540 93 Arg Val Glu Asp Phe Gly Asn Ala Trp Lys Leu His Gly Asp Cys Gln 550 555 95 Asp Leu Gln Lys Gln His Ser Asp Pro Cys Ala Leu Asn Pro Arg Met 565 570 97 Thr Arg Phe Ser Glu Glu Ala Cys Ala Val Leu Thr Ser Pro Thr Phe 585 99 Glu Ala Cys His Arg Ala Val Ser Pro Leu Pro Tyr Leu Arg Asn Cys 595 600 101 Arg Tyr Asp Val Cys Ser Cys Ser Asp Gly Arg Glu Cys Leu Cys Gly RAW SEQUENCE LISTING DATE: 08/04/2006
PATENT APPLICATION: US/10/531,427 TIME: 13:11:47

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102		610					615					620				
			λla	Sar	Тугу	בות		λla	Cvc	Nlα	Clar		C1.,	Val	7.~~	17-1
	625	DCu	niu	JCI	- 7 -	630	лια	AIG	Cys	AIG	635	Arg	Gry	vai	Arg	640
		Trn	Δτα	Glu	Dro		λνα	Cvc	Glu	Lou		Caro	Dro	Lys	C1	
106	nια	TIP	nr 9	OIU	645	Gry	nr 9	Cys	Giu	650	ASII	Cys	FIO	пуъ	655	GIII
	Va 1	Tarr	T.011	Gln		Glv	Thr	Dro	Cvc		Lou	Thr	Cvc	Arg		T 011
108	val	тут	цец	660	Cys	Gry	1111	PIO	665	ASII	шец	1111	Cys	670	ser	Бец
	Sar	ጥኒኒ	Dro		Glu	Glu	Cvc	λcn		λla	Cara	T 011	C1.,	Gly	Crra	Dho
110	DCI	ıyı	675	Asp	GIU	Giu	Cys	680	GIU	нта	Cys	Deu	685	GIY	Cys	Pije
	Cve	Pro		Glv	T.611	ጥኒም	Mot		Glu	λνα	Glv	λαn		Val	Dro	Lvc
112	Cyb	690	110	O _T y	ДСu	171	695	АЗР	Giu	nr 9	Gry	700	Cys	Val	FIO	цуз
	Δla		Cvs	Pro	Cvs	Tvr		Asn	Glv	Glu	Tle		Gln	Pro	Glu	Δen
	705	01	Cy D	110	Cyb	710	- 7 -	nop	OLY	OIU	715	1110	GIII	110	GIU	720
		Phe	Ser	Δsn	Hic	_	Thr	Met	Cve	Туг		G111	Acn	Gly	Dha	
116			JCI	nop	725	1115	1111	ricc	Cys	730	Cys	Giu	ьэр	Gry	735	Mec
	His	Cvs	Thr	Met		Glv	Val	Pro	Glv		T.e.11	T.e.11	Pro	Asp		Wa 1
118		C ₁ C	****	740	DCI	Ory	V 41	110	745	UCI	Dea	пси	110	750	nia	vai
	Len	Ser	Ser		T.e.11	Ser	His	Ara		Tare	Ara	Ser	T.611	Ser	Cvc	Δrσ
120			755			501		760	561	2,5	**** 9	501	765	001	Cys	n-9
	Pro	Pro		Val	Lvs	Leu	Val		Pro	Ala	Asn	Asn		Arg	Δla	Glu
122		770			_, _	200	775	cy 5	110	111 U	тор	780	LCu	g	nia	Olu
	Glv	_	Glu	Cvs	Thr	Ivs		Cvs	Gln	Asn	Tvr		Len	Glu	Cvs	Met
	785			-1-		790		-1-			795			024	0,70	800
		Met	Glv	Cvs	Val	Ser	Glv	Cvs	Leu	Cvs		Pro	Glv	Met	Val	
126			1	-1-	805		1	-7-		810			0- 1		815	
	His	Glu	Asn	Arg	Cvs	Val	Ala	Leu	Glu		Cvs	Pro	Cvs	Phe		Gln
128				820	•				825		- 2		-2-	830		
129	Gly	Lys	Glu	Tyr	Ala	Pro	Glv	Glu		Val	Lvs	Ile	Glv	Cys	Asn	Thr
130	-	•	835	•			1	840			•		845	_		
131	Cys	Val	Cys	Arg	Asp	Arq	Lys	Trp	Asn	Cys	Thr	Asp	His	Val	Cys	qaA
132	-	850	_	_	_		855	_		-		860			•	-
133	Ala	Thr	Cys	Ser	Thr	Ile	Gly	Met	Ala	His	Tyr	Leu	Thr	Phe	Asp	Gly
	865					870	_				875				_	880
135	Leu	Lys	Tyr	Leu	Phe	Pro	Gly	Glu	Cys	Gln	Tyr	Val	Leu	Val	Gln	Asp
136					885					890					895	
137	Tyr	Cys	Gly	Ser	Asn	Pro	Gly	Thr	Phe	Arg	Ile	Leu	Val	Gly	Asn	Lys
138				900					905			Ū		910		
139	Gly	Cys	Ser	His	Pro	Ser	Val	Lys	Cys	Lys	Lys	Arg	Val	Thr	Ile	Leu
140			915					920					925			
141	Val	Glu	Gly	Gly	Glu	Ile	Glu	Leu	Phe	Asp	Gly	Glu	Val	Asn	Val	Lys
142		930					935					940				
143	Arg	Pro	Met	Lys	Asp	Glu	Thr	His	Phe	Glu	Val	Val	Glu	Ser	Gly	Arg
	945					950					955					960
145	Tyr	Ile	Ile	Leu	Leu	Leu	Gly	Lys	Ala	Leu	Ser	Val	Val	Trp	Asp	Arg
146					965					970					975	
	His	Leu	Ser		Ser	Val	Val	Leu		Gln	Thr	Tyr	Gln	Glu	Lys	Val
148		_		980					985					990		
	Cys	Gly		Cys	Gly	Asn		_	Gly	Ile	Gln			Asp	Leu	Thr
150			995				1	1000				1	1005			

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151 Ser Ser Asn Leu Gln Val Glu Glu Asp Pro Val Asp Phe Gly Asn Ser 1010 1015 153 Trp Lys Val Ser Ser Gln Cys Ala Asp Thr Arg Lys Val Pro Leu Asp 1030 154 1025 1035 155 Ser Ser Pro Ala Thr Cys His Asn Asn Ile Met Lys Gln Thr Met Val 156 1045 1050 157 Asp Ser Ser Cys Arg Ile Leu Thr Ser Asp Val Phe Gln Asp Cys Asn 158 1060 1065 159 Lys Leu Val Asp Pro Glu Pro Tyr Leu Asp Val Cys Ile Tyr Asp Thr 160 1075 1080 161 Cys Ser Cys Glu Ser Ile Gly Asp Cys Ala Cys Phe Cys Asp Thr Ile 162 1090 1100 1095 163 Ala Ala Tyr Ala His Val Cys Ala Gln His Gly Lys Val Val Thr Trp 1110 1115 165 Arg Thr Ala Thr Leu Cys Pro Gln Ser Cys Glu Glu Arg Asn Leu Arg 1125 1130 167 Glu Asn Gly Tyr Glu Cys Glu Trp Arg Tyr Asn Ser Cys Ala Pro Ala 1140 1145 169 Cys Gln Val Thr Cys Gln His Pro Glu Pro Leu Ala Cys Pro Val Gln 1155 1160 1165 171 Cys Val Glu Gly Cys His Ala His Cys Pro Pro Gly Lys Ile Leu Asp 1175 1180 173 Glu Leu Leu Gln Thr Cys Val Asp Pro Glu Asp Cys Pro Val Cys Glu 174 1185 1190 1195 175 Val Ala Gly Arg Arg Phe Ala Ser Gly Lys Lys Val Thr Leu Asn Pro 1205 1210 1215 177 Ser Asp Pro Glu His Cys Gln Ile Cys His Cys Asp Val Val Asn Leu 178 1220 1225 179 Thr Cys Glu Ala Cys Gln Glu Pro Gly Gly Leu Val Val Pro Pro Thr 1240 1245 181 Asp Ala Pro Val Ser Pro Thr Thr Leu Tyr Val Glu Asp Ile Ser Glu 1250 1255 183 Pro Pro Leu His Asp Phe Tyr Cys Ser Arg Leu Leu Asp Leu Val Phe 1270 1275 185 Leu Leu Asp Gly Ser Ser Arg Leu Ser Glu Ala Glu Phe Glu Val Leu 1285 1290 187 Lys Ala Phe Val Val Asp Met Met Glu Arg Leu Arg Ile Ser Gln Lys 1300 1305 189 Trp Val Arg Val Ala Val Val Glu Tyr His Asp Gly Ser His Ala Tyr 190 1315 1320 1325 191 Ile Gly Leu Lys Asp Arg Lys Arg Pro Ser Glu Leu Arg Arg Ile Ala 192 1330 1335 1340 193 Ser Gln Val Lys Tyr Ala Gly Ser Gln Val Ala Ser Thr Ser Glu Val 1350 1355 195 Leu Lys Tyr Thr Leu Phe Gln Ile Phe Ser Lys Ile Asp Arg Pro Glu 1365 1370 197 Ala Ser Arg Ile Ala Leu Leu Met Ala Ser Gln Glu Pro Gln Arg 1380 1385 199 Met Ser Arg Asn Phe Val Arg Tyr Val Gln Gly Leu Lys Lys Lys

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1395 1400 201 Val Ile Val Ile Pro Val Gly Ile Gly Pro His Ala Asn Leu Lys Gln 1410 1415 203 Ile Arg Leu Ile Glu Lys Gln Ala Pro Glu Asn Lys Ala Phe Val Leu 1430 1435 205 Ser Ser Val Asp Glu Leu Glu Gln Gln Arg Asp Glu Ile Val Ser Tyr 1445 1450 207 Leu Cys Asp Leu Ala Pro Glu Ala Pro Pro Pro Thr Leu Pro Pro His 208 1460 1465 209 Met Ala Gln Val Thr Val Gly Pro Gly Leu Leu Gly Val Ser Thr Leu 210 1475 1480 1485 211 Gly Pro Lys Arg Asn Ser Met Val Leu Asp Val Ala Phe Val Leu Glu 212 1490 1495 1500 213 Gly Ser Asp Lys Ile Gly Glu Ala Asp Phe Asn Arg Ser Lys Glu Phe 214 1505 1510 1515 215 Met Glu Glu Val Ile Gln Arg Met Asp Val Gly Gln Asp Ser Ile His 1525 . 1530 217 Val Thr Val Leu Gln Tyr Ser Tyr Met Val Thr Val Glu Tyr Pro Phe 218 . 1540 1545 1550 219 Ser Glu Ala Gln Ser Lys Gly Asp Ile Leu Gln Arg Val Arg Glu Ile 220 1555 1560 221 Arg Tyr Gln Gly Gly Asn Arg Thr Asn Thr Gly Leu Ala Leu Arg Tyr 222 1570 1575 1580 223 Leu Ser Asp His Ser Phe Leu Val Ser Gln Gly Asp Arg Glu Gln Ala 224 1585 1590 1595 225 Pro Asn Leu Val Tyr Met Val Thr Gly Asn Pro Ala Ser Asp Glu Ile 1605 1610 1615 227 Lys Arg Leu Pro Gly Asp Ile Gln Val Val Pro Ile Gly Val Gly Pro 228 1620 1625 229 Asn Ala Asn Val Gln Glu Leu Glu Arg Ile Gly Trp Pro Asn Ala Pro 1635 1640 231 Ile Leu Ile Gln Asp Phe Glu Thr Leu Pro Arg Glu Ala Pro Asp Leu 1655 1660 233 Val Leu Gln Arg Cys Cys Ser Gly Glu Gly Leu Gln Ile Pro Thr Leu 1670 1675 235 Ser Pro Ala Pro Asp Cys Ser Gln Pro Leu Asp Val Ile Leu Leu Leu 1685 1690 237 Asp Gly Ser Ser Ser Phe Pro Ala Ser Tyr Phe Asp Glu Met Lys Ser 238 1700 1705 239 Phe Ala Lys Ala Phe Ile Ser Lys Ala Asn Ile Gly Pro Arg Leu Thr 240 1715 1720 1725 241 Gln Val Ser Val Leu Gln Tyr Gly Ser Ile Thr Thr Ile Asp Val Pro 242 1730 1735 1740 243 Trp Asn Val Val Pro Glu Lys Ala His Leu Leu Ser Leu Val Asp Val 1750 1755 245 Met Gln Arg Glu Gly Gly Pro Ser Gln Ile Gly Asp Ala Leu Gly Phe 1765 1770 247 Ala Val Arg Tyr Leu Thr Ser Glu Met His Gly Ala Arg Pro Gly Ala 1780 1785

RAW SEQUENCE LISTING ERROR SUMMARY DATE: 08/04/2006 PATENT APPLICATION: US/10/531,427 TIME: 13:11:49

Input Set : A:\2005-11-10 0020-5363PUS1.txt
Output Set: N:\CRF4\08042006\J531427.raw

Invalid Line Length:

The rules require that a line not exceed 72 characters in length. This includes spaces.

Seq#:1; Line(s) 6
Seq#:7; Line(s) 493
Seq#:8; Line(s) 504
Seq#:9; Line(s) 515
Seq#:10; Line(s) 526
Seq#:11; Line(s) 537
Seq#:12; Line(s) 548

VERIFICATION SUMMARY

DATE: 08/04/2006

PATENT APPLICATION: US/10/531,427

TIME: 13:11:49

Input Set : A:\2005-11-10 0020-5363PUS1.txt Output Set: N:\CRF4\08042006\J531427.raw